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Recurrent bowel infarction in paroxysmal nocturnal haemoglobinuria

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In paroxysmal nocturnal haemoglobinuria (PNH), thrombotic complications are a common cause of death. Abdominal pain requires particularly careful assessment.

CASE HISTORY

A man of 43 was referred after 24 hours of feeling generally unwell with intermittent rigors. PNH had been diagnosed 18 years previously, and regular blood transfusions for his anaemia had resulted in iron-overload diabetes mellitus. 8 years before the present episode he had had a small-bowel resection for ischaemia secondary to venous thrombosis.

On examination his abdomen was distended and slightly tender in the left lower quadrant, without obvious peritonism. Haemoglobin was 6.4 g/dL and platelet count 16×10^9/L. Blood indices were otherwise normal. A plain abdominal X-ray showed dilated small-bowel loops. Adhesional small-bowel obstruction was diagnosed and initial management was conservative. After three days the abdominal distention had not resolved and a CT scan (Figure 1) showed gross small-bowel dilatation with an area of collapsed small bowel distally, highly suggestive of a mechanical obstruction. At laparotomy, with blood-transfusion and platelet cover, he was found to have a segment of gangrenous small bowel with a stricture. This was resected and the bowel was anastomosed by an extramucosal technique. The patient spent the first three postoperative days on the intensive care unit for close monitoring, and received total parenteral nutrition for five days until enteral feeding was started. No further blood or platelet transfusions were required. Histopathological examination of the operative specimen showed extensive haemorrhagic infarction secondary to venous thrombosis.

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COMMENT

PNH is an acquired disease characterized by attacks of intravascular haemolysis and haemoglobinuria. It can lead to profound anaemia, thrombocytopenia and leucopenia and is often complicated by venous thrombosis.[1] The thrombotic tendency may be related to deficiency of a glycosyl-phosphatidylinositol anchor protein.[2] Thrombotic complications are the most common cause of death in PNH, and about 40% of patients have one or more episodes of venous thrombosis during their illness.[3] The incidence of mesenteric venous thrombosis leading to bowel ischaemia is between 3% and 8%.[4]

In 1966, Blum and Gardner[5] described successful laparotomy in two patients with PNH—one with infarction of the terminal ileum, who underwent segmental resection; the other with an infarcted caecum, treated with a limited right hemicolectomy. Subsequently, Doukas *et al*.[6] reported resection of two areas of ischaemic bowel in another patient, with good postoperative results. A fourth case, a man of 37 who survived after small-bowel resection for bowel gangrene secondary to extensive venous thrombosis, was reported by Williamson *et al*.[7] in 1987. These were the only reports we could find of patients with PNH surviving

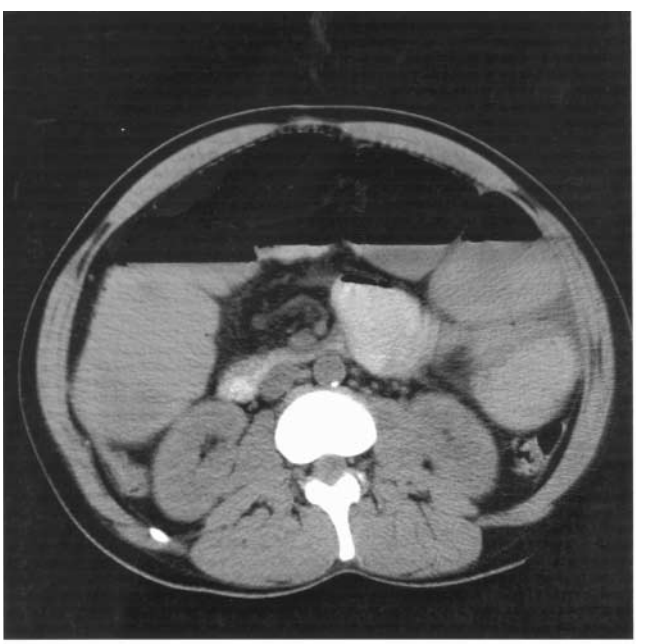

Figure 1 **CT scan of abdomen on day 3**

laparotomy for bowel ischaemia. None had survived more than one laparotomy for this condition, as ours did.

A difficulty in dealing with these cases is that abdominal pain is a frequent symptom in PNH and is seldom due to bowel thrombosis. The possibility simply has to be borne in mind. In some cases, conservative management with intravenous fluids, antibiotics and low-molecular-weight heparin is successful.[8]

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Priapism at age 94

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Priapism is defined as persistent penile erection lasting beyond 6 hours. 50–60% of cases are idiopathic but known causes include drugs, sickle-cell anaemia, leukaemia, pelvic tumours, spinal cord injury and trauma.

CASE HISTORY

A man aged 94 was admitted after three days of increasing lower limb pain, worse on the left than the right. In addition, priapism had developed. He had type 2 diabetes and there was a history of heavy smoking. 10 years previously an abdominal aortic aneurysm had been repaired with a straight inlay graft. On examination he had an ill-defined non-tender pulsatile fullness at the level of the umbilicus and a moderately turgid erection. His legs were bluish and mottled with slight oedema on the left side. No pulses were palpable below the femoral arteries, but pulsation was felt along the proximal greater saphenous veins bilaterally. Doppler studies identified distal pulses bilaterally and confirmed arterial pulsation in the saphenous veins. CT scans (Figures 1 and 2) revealed contrast enhancement of the inferior vena cava (IVC) during the arterial phase and also a pseudoaneurysm, related to the distal anastomosis of his aortic graft, through which contrast was seen running into the IVC. The patient was anuric and

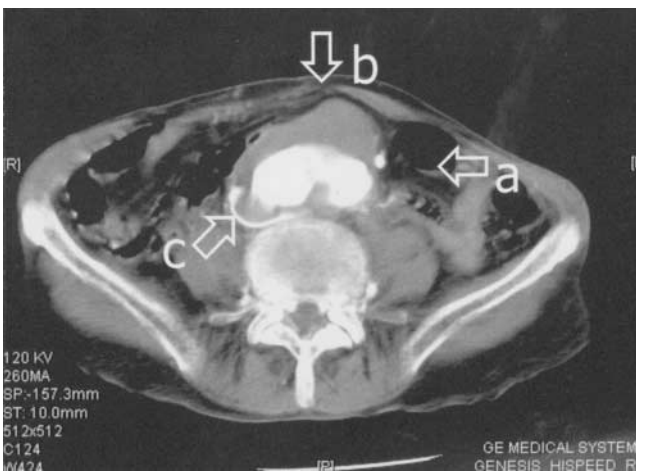

Figure 1 **Pseudoaneurysm around bifurcation of the aorta just below lower anastomotic line.** Note flow of contrast into vena cava around area of turbulence or thrombosis. a=aortic bifurcation just distal to inlay graft anastomosis; b=anastomotic pseudoaneurysm; c=contrast flowing into vena cava during arterial phase (effect diluted by turbulence or possible thrombosis)

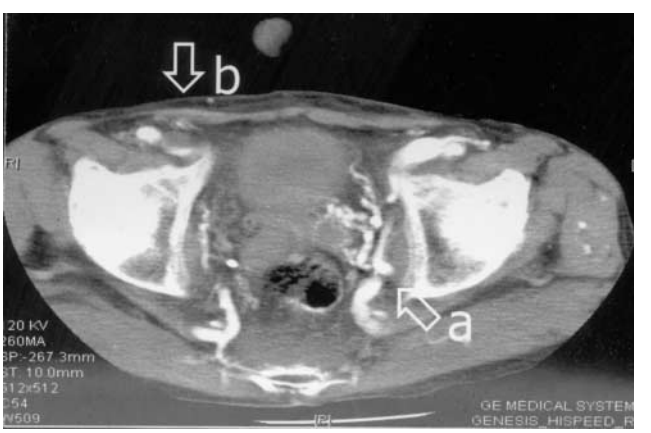

Figure 2 **Arterial phase congestion of pelvic vessels and left femoral vein.** Note right femoral vein thrombosis. a=congested pelvic veins during arterial phase; b=thrombosis in right femoral vein

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